Case Report

Unusual case of infantile fibrosarcoma evaluated on F-18 fluorodeoxyglucose positron emission tomography-computed tomography

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ABSTRACT

Infantile fibrosarcoma (IFS) is a rare soft-tissue sarcoma originating from extremities and occasionally from axial soft tissue. The prognosis is good with favorable long-term survival. It is rarely metastasizing tumor, the chances being lesser with IFS originating from extremities. Use of neoadjuvant chemotherapy (NACT) as a treatment regime further reduces the chances of local relapse and distant metastasis. The organs commonly affected in metastatic IFS are lungs and lymph nodes. We report an unusual case of an IFS originating from extremity, which received NACT, yet presented with an early metastatic disease involving soft tissues and sparing lungs and lymph nodes, as demonstrated on fluorodeoxyglucose positron emission tomography-computed tomography.

Keywords: Fluorodeoxyglucose positron emission tomography-computed tomography, infantile fibrosarcoma, unusual metastasis

INTRODUCTION

Infantile fibrosarcoma (IFS) is an uncommon neoplasm. Pediatric soft-tissue sarcomas (STSs) account for approximately 7% of all childhood tumors.[1] out of which only 5–10% are IFSs.[2] IFS can originate more commonly from extremities.

IFS is locally aggressive neoplasm which rarely metastasizes. The distant metastatic sites are most commonly lungs and nodes. There is satisfactory prognosis in terms of survival after curative treatment.[3] Though surgery is the mainstay of treatment, advent of neoadjuvant chemotherapy (NACT) has reduced the incidence of recurrence/metastasis.[4]

We present a case of IFS with early and unusual distant metastasis, localized on fluorodeoxyglucose positron emission tomography-computed tomography (FDG PET-CT) scan. It has discriminating power in evaluating N and M stages of STSs.

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CASE REPORT

The patient, a 4-year-old girl child, is a known case of IFS, primarily involving right proximal tibia and staging whole body PET/magnetic resonance (MR) scan not suggestive of disease elsewhere. She received NACT followed by wide local excision. She was detected with local relapse 3 months later for which she underwent chemotherapy followed by right above knee amputation. Histopathology was suggestive of residual viable IFS with clear resection margins. Complaint of right thigh pain originated 10 months after the amputation.

On physical examination, she had a palpable mildly tender lump in right proximal thigh. An MR imaging of the right thigh was obtained, which showed a neoplastic lesion involving right proximal shaft of femur. The patient was referred for FDG PET-CT.

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The maximum intensity projection [Figure 1] was suggestive of significant tracer uptake in right facial, lower lumbosacral, and bilateral proximal thigh regions. The axial CT and fused PET-CT images demonstrated FDG avid hypermetabolic foci in soft-tissue lesions involving right zygoma with intraorbital extension (maximum standardized uptake value [SUVmax 9]) [Figure 2a], right proximal femur [SUVmax 6.1, Figure 2b], sacrum with involvement of right sacroiliac joint [SUVmax 16, Figure 2c], and right suprascapular region [SUVmax 9, Figure 2d].

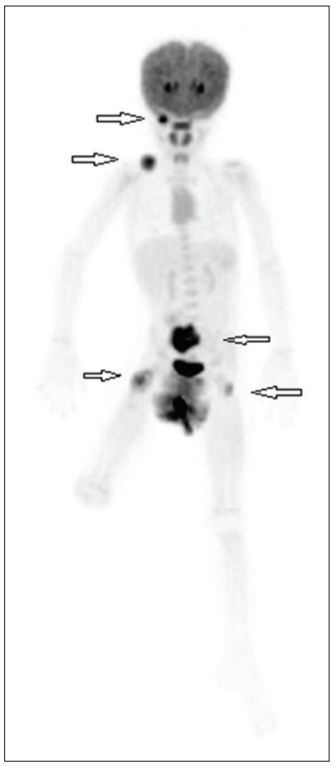

Figure 1: Maximum intensity projection of F-18 fluorodeoxyglucose positron emission tomography-computed tomography showing increased tracer uptake in regions pointed by arrowheads

Subsequently, biopsy was obtained from sacral and right proximal femoral lesions. The tumor cells were arranged in interlacing fascicles and herringbone pattern [Figure 3a], with pleomorphic nuclei and brisk mitotic activity of >10/10 hpf. Many blood vessels were visualized [Figure 3b]. It was suggestive of viable high-grade IFS. Immunohistochemistry showed high MIB-1 labeling index.

DISCUSSION

IFS has incidence of 5 per million in the pediatric age group and majority of the patients are <2 years old.[5] Although its etiology is unknown, a fusion transcript ETV6-NTRK3 resulting from a chromosomal translocation t(12;15) is commonly associated.[3] The extremities account for the primary in almost 74% of the cases, followed by head and neck region.[6] Most common site of origin is around knee joint followed by thigh and hip regions.

The pathologic appearance of IFS includes spindle cells with many blood vessels arranged in hemangiopericytoma-like fascicular pattern.[7]

The primary modality of treatment is surgery. Radical surgery performed to achieve tumour free margins has higher morbidity.[5,6] Advent of NACT allowed conservative surgeries and decreased local as well as metastatic relapses. In a large study comprising 56 infants by Orbach *et al*., 20 patients were given initial courses of chemotherapy before surgery. Seventy-three percent of these were found to be chemosensitive and 22% maintained stable disease. Only 1 patient (5%) developed distant metastasis.[4]

IFS has favorable survival. Akyüz *et al*. performed a retrospective study with 11 patients who were treated with surgery alone or NACT + surgery. Of those, 2 patients died due to chemotherapy-related complications. One patient died who had metastatic disease to lungs and brain at presentation itself. Rest, all

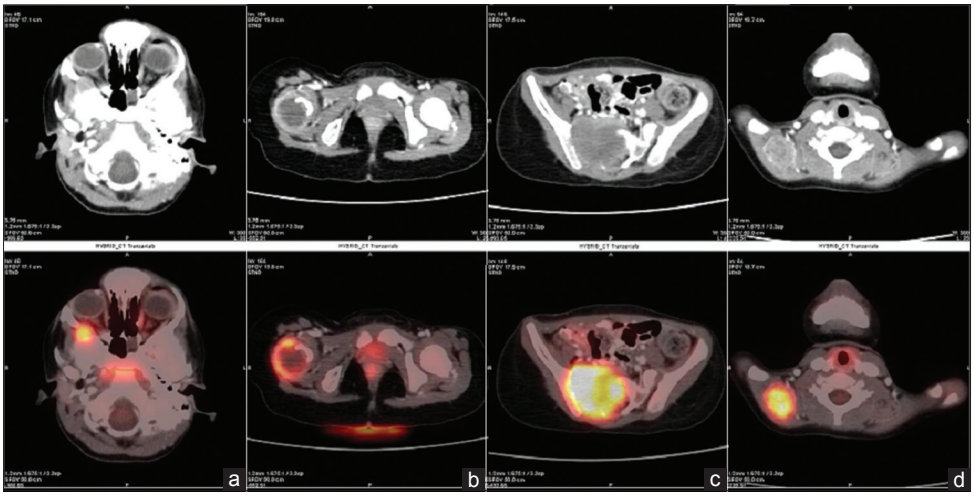

Figure 2: Transaxial computed tomography and fusion positron emission tomography-computed tomography images on F-18 fluorodeoxyglucose positron emission tomography-computed tomography scan showing fluorodeoxyglucose avid hypermetabolic foci in soft-tissue lesions involving; (a) right zygomatic bone with intraorbital extension, (b) right proximal femur, (c) sacrum and right sacroiliac joint, and (d) right suprascapular region

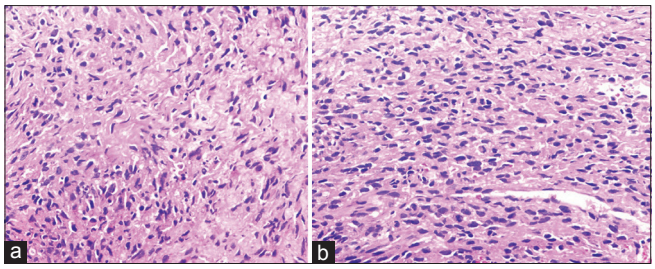

Figure 3: Microscopic images of hematoxylin and eosin stained biopsy slides obtained from sacral and right proximal femur lesions showing; (a) arrangement of tumor cells in interlacing fascicles and herringbone pattern, with (b) multiple blood vessels dispersed within the tumor tissue

were followed up for varying periods of 18 months to 13.5 years and were found to be disease free.[5] Another study considered 4 patients of IFS who underwent NACT + wide local excision. Three patients who had primary in extremities achieved complete remission and were disease free even after 2 years of follow-up. One infant had a neck primary and developed metastases to brain, lungs, and kidneys, which responded to chemotherapy.[8] In addition, studies have proved that IFS carries overall 5-year survival rate of >85%.[3,4,6]

Overall, 17–43% of cases present with local recurrence within 1 year after curative treatment.[6] The incidence of distant metastases was found to be 5–10%. Further, IFS originating from extremities is found less likely to cause distant metastases (8%) than axial IFS (26%).[5]

WHO classification of STSs categorizes IFS in intermediate rarely metastasizing neoplasm, which is defined as the ones which have a documented ability to metastasize but seldom do. As a group, they have <2% chances of distant spread which cannot be predicted on the basis of histomorphology.[9] The more common sites of distant metastasis are lungs and lymph nodes although distant seeding of kidneys and brain have been documented.[5,8]

FDG PET-CT is established modality in assessment of malignancies. It has been well documented in adult STSs for staging, response evaluation, and recurrence setting. In a meta-analysis, the sensitivity and specificity of FDG PET-CT in identifying primary as well as metastatic disease in STSs were found to be 92% and 73%.[10] Chen *et al*. reported a case of IFS with local recurrence demonstrated by FDG PET-CT. The lesion was found to be low-grade FDG avid (SUVmax <2.5).[6]

CONCLUSION

IFS is rare, soft-tissue neoplasm of rarely metastasizing potential. Posttreatment prognosis is good in terms of disease-free survival. Lungs, nodes, and brain were the common site of metastasis. Our case of this rare pediatric neoplasm originating from an extremity uniquely presented with distant soft-tissue metastasis within 1 year of receiving NACT and surgery. The FDG avidity of metastatic lesions was unusually high unlike reported in the literature so far. Our findings revealed FDG PET-CT as a potentially useful tool in detection of metastatic disease recurrence in IFS cases. Furthermore, metabolic parameters such as SUVmax can be evaluated as prognostic tools to assess the tumor aggressiveness.

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Conflicts of interest
There are no conflicts of interest.

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